

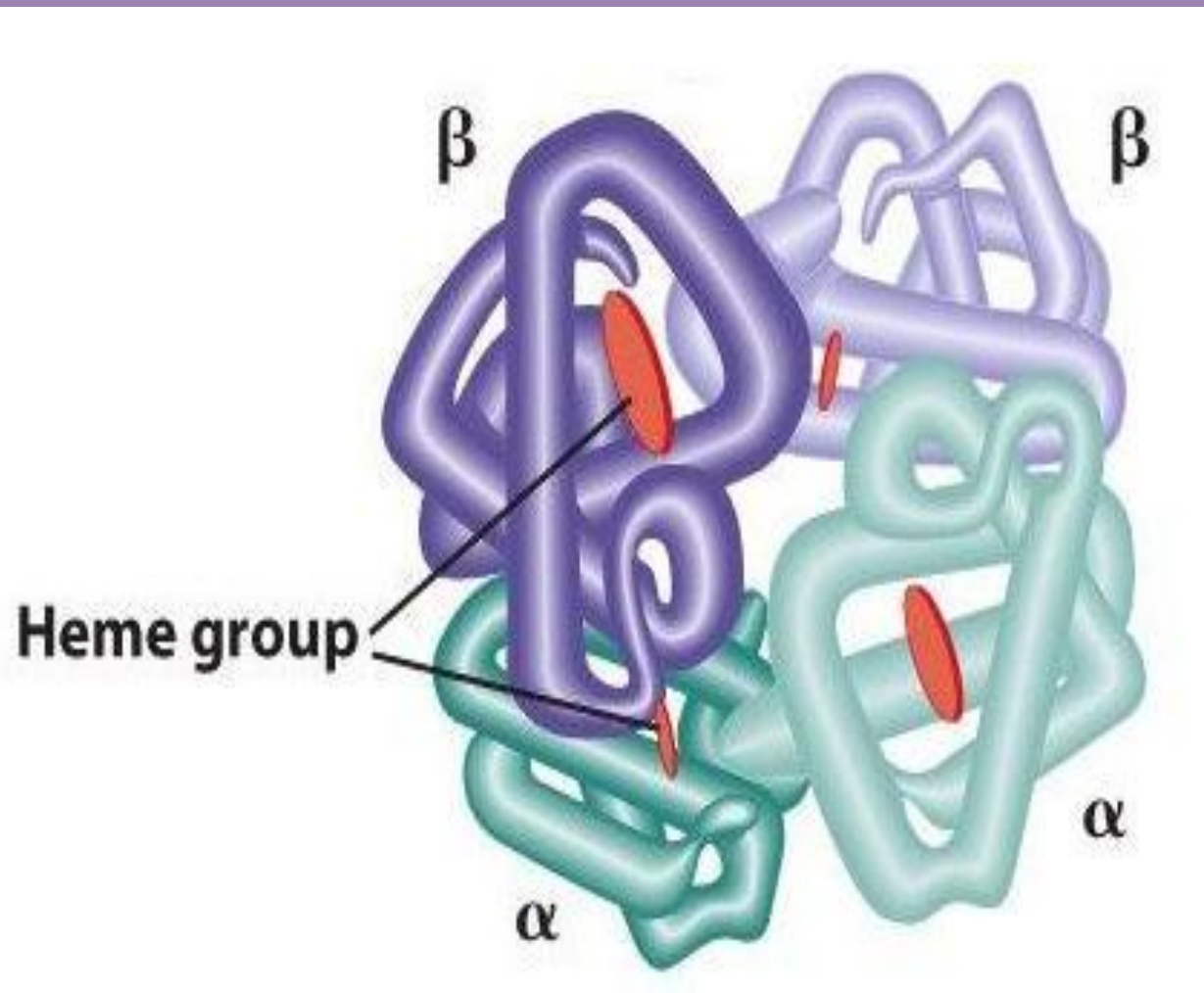
# Disorders of Red Blood Cells

*Produced by: Assistant Professor Dr. Ala'a Hassan Mirza Hussain*

The function of RBC, facilitated by hemoglobin molecule, is to transport oxygen to the tissue.

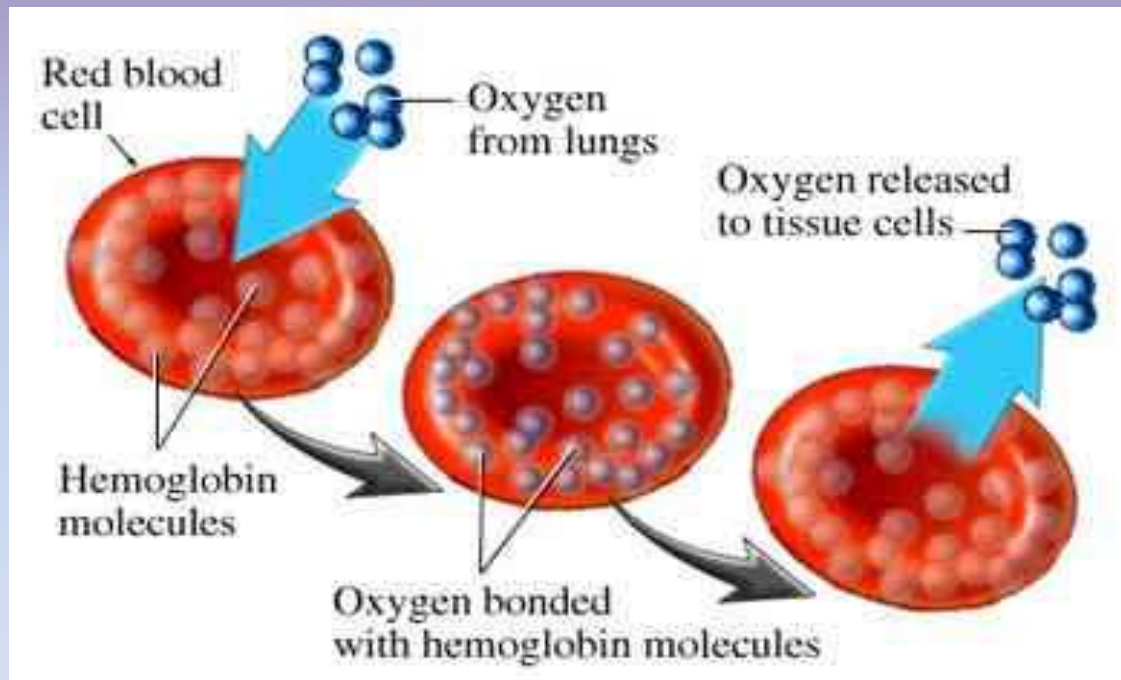
Oxygen is poorly soluble in plasma, So about 95%-98% is carried bound to hemoglobin.

The Hb molecule is composed of two pairs of structurally different  $\alpha$  and  $\beta$  chains. Each of four polypeptide chains consists of globin (protein) and a haeme unit which surrounds an atom of iron that binds oxygen. Thus Hb can carry four molecules of Oxygen.



- The production of each type of globin chain is controlled by individual structural genes with five different gene loci.
- Mutations which occurs in anywhere in these five loci resulted over 550 types of abnormal Hb molecules.

# Red cell Metabolism



- The RBC lacks mitochondria; it depends on glucose and glycolytic pathway for its metabolic needs.
- The enzyme mediated anaerobic metabolism of glucose generates the ATP need for normal membrane function and ion transport.
- The depletion of glucose or the functional deficiency of one the glycolytic enzymes leads to the premature death of RBC.

An offshoot of the glycolytic pathway is the production of **2,3-diphosphoglycerate (2,3-DPG)**, which binds to the Hb molecule and reduces the affinity of Hb for Oxygen.

This facilitates the release of Oxygen at the tissue level.

- An increase in the concentration of 2,3- DPG occurs in conditions:

Chronic hypoxia like in case anemia, chronic lung disease, and residence at high attitudes.

# Hemoglobin Oxidation

- The combination of Hb with oxygen can be interrupted by certain chemicals (e.g. nitrates and sulfates) and drugs that oxidize Hb to the inactive form.
- The nitrite ion reacts with Hb to produce methemoglobin, which has a low affinity for Oxygen.
- Large doses of nitrites can result in high levels of methemoglobin, causing pseudocyanosis and tissue hypoxia. For example, sodium nitrate, which is used in curing meat, can produce methemoglobin when taken in large amounts.



# Anemia

- Abnormally low number of circulating RBC or level of Hb or both, resulting in diminished oxygen-carrying capacity.
- Causing:
  - 1). Excessive loss of RBC:
    - Hemorrhage
    - Hemolysis
  - 2). Impaired RBC production result from:
    - Depression in bone marrow
    - Inhered
    - Nutritional
    - Bone marrow cancer

# Hemolytic Anemia

- It is characterized by the premature destruction of RBC , the retention in the body of iron and the other products of Hb destruction and an increase in erythropoiesis.
- All most all types of hemolytic anemia are distinguished by normocytic normochromic RBC

- Because of the red blood cells shortened life span, the bone marrow usually is hyperactive, resulting an increased number of reticulocytes in the circulating blood.
- There is hemoglobinuria and hemoglobinemia and jaundice when the hemolysis occurs intravascular.
- The person is easy fatigable, dyspnea, tachycardia